

**Amendment**  
**in the Specification**

Please replace Table 1 beginning on page 21, line 24 with the following Table 1.

**Table 1**

**Localization Signals for Targeting to the Mitochondria.**  
 (verified using Mitochondrial Project MITOP Database --  
<http://mips.gsf.de/proj/medgen/mitop/>)

MITOP Designation	SEQ. ID. NO.	Gene Name	Gene Name Full
106092	7	Etfa	electron transfer flavoprotein alpha chain precursor – mouse (SEQ ID NO. 7)
106098	8	Etfb	electron transfer flavoprotein beta chain – mouse (SEQ ID NO. 8)
107450	9	Dld	dihydrolipoamide dehydrogenase precursor – human (SEQ ID NO. 9)
87979	10	Ak3	nucleoside-triphosphate--adenylate kinase 3 – mouse (SEQ ID NO. 10)
88529	11	Cs	citrate synthase, mitochondrial (SEQ ID NO. 11)
891996	12	Cps1	carbamoyl-phosphate synthetase 1 (SEQ ID NO. 12)
97045	13	Mod2	malic enzyme complex, mitochondrial – mouse (SEQ ID NO. 13)
97499	14	Pcca	propionyl-CoA carboxylase alpha chain precursor – mouse (SEQ ID NO. 14)
A27883	15	PCCA	propionyl-CoA carboxylase alpha chain precursor (SEQ ID NO. 15)
A28053	16	Cbr2	carbonyl reductase (NADPH) – mouse (SEQ ID NO. 16)
A29881	17	mpp-2	Mitochondrial processing peptidase beta subunit precursor (beta-mpp) (ubiquinol-cytochrome c reductase complex core protein I) (SEQ ID NO. 17)
A30605	18	ACADS	acyl-CoA dehydrogenase precursor, short-chain-specific (SEQ ID NO. 18)
A31998	19	ETFA	electron transfer flavoprotein alpha

MITOP Designation	SEQ. ID. NO.	Gene Name	Gene Name Full
			chain precursor (SEQ ID NO. 19)
A32422	20	DBT	Dihydrolipoamide S-(2-methylpropanoyl)transferase precursor (SEQ ID NO. 20)
A32800	21	HSPD1	heat shock protein 60 precursor (SEQ ID NO. 21)
A36442	22	mpp-1	Mitochondrial processing peptidase alpha chain precursor (SEQ ID NO. 22)
A37033	23	IVD	isovaleryl-CoA dehydrogenase precursor (SEQ ID NO. 23)
A37157	24	BCKD	3-methyl-2-oxobutanoate dehydrogenase (lipoamide) E1-beta chain precursor (SEQ ID NO. 24)
A38234	25	OGDH	Oxoglutarate dehydrogenase (lipoamide) precursor (SEQ ID NO. 25)
A39503	26	ME2	Malate dehydrogenase (NAD <sup>+</sup> ) precursor, mitochondrial (SEQ ID NO. 26)
A40487	27	FDXR	ferredoxin--NADP <sup>+</sup> reductase, long form, precursor (SEQ ID NO. 27)
A40559	28	ACADL	long-chain-acyl-CoA dehydrogenase (LCAD) (SEQ ID NO. 28)
A40872	29	ALDH5	aldehyde dehydrogenase (NAD <sup>+</sup> ) 5 precursor, mitochondrial (SEQ ID NO. 29)
A41581	30	CYP3	peptidylprolyl isomerase 3 precursor (SEQ ID NO. 30)
A42224	31	arg-2	Carbamoyl-phosphate synthase, arginine-specific, small chain precursor (arginine-specific carbamoyl-phosphate synthetase, glutamine chain) (cps-a) (SEQ ID NO. 31)
A42845	32	BDH	D-beta-hydroxybutyrate dehydrogenase precursor (3-hydroxybutyrate dehydrogenase) (fragment) (SEQ ID NO. 32)
A45470	33	HMGC	Hydroxymethylglutaryl-CoA lyase (SEQ ID NO. 33)

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A47255	34	Pcx	pyruvate carboxylase (SEQ ID NO. 34)
A53020	35	PCCB	propionyl-CoA carboxylase beta chain precursor (SEQ ID NO. 35)
A53719	36	GLUDP	glutamate dehydrogenase (NAD(P)+) 2 precursor (SEQ ID NO. 36)
A55075	37	HspE1	chaperonin-10 (SEQ ID NO. 37)
A55680	38	ACADS	short/branched chain acyl-CoA dehydrogenase precursor (SEQ ID NO. 38)
A55723	39	DCI	dodecenoyl-CoA Delta-isomerase precursor, mitochondrial (SEQ ID NO. 39)
A55724	40	Acadm	Acyl-CoA dehydrogenase, medium-chain specific precursor (MCAD) (SEQ ID NO. 40)
AA227572	41	WARS2	tryptophanyl-tRNA synthetase 2 (mitochondrial) – human (SEQ ID NO. 41)
AB029948	42	SerRS	mitochondrial seryl-tRNA synthetase (cDNA FLJ20450 FIS, CLONE KAT05607) – human (SEQ ID NO. 42)
ACDL_MOUSE	43	Acadl	Acyl-CoA dehydrogenase, long-chain specific precursor (LCAD) (SEQ ID NO. 43)
AF047042	44	CS	citrate synthase, mitochondrial (SEQ ID NO. 44)
AF097441	45	FARS1	phenylalanine-tRNA synthetase (FARS1) mRNA, nuclear gene encoding mitochondrial protein – human (SEQ ID NO. 45)
ATPO_HUMAN	46	ATP5O	ATP synthase oligomycin sensitivity conferral protein precursor, mitochondrial (SEQ ID NO. 46)
AXHU	47	FDX1	adrenodoxin precursor (SEQ ID NO. 47)
CCHU	48	HCS	cytochrome c (SEQ ID NO. 48)
CCNC	49	cyc-1	Cytochrome c (SEQ ID NO. 49)
CE06620	50	-	Probable leucyl-tRNA synthetase, mitochondrial (SEQ ID NO. 50)
CE09597	51	-	Pyruvate dehydrogenase (E2) dihydrolipoamide acetyltransferase

MITOP Designation	SEQ. ID. NO.	Gene Name	Gene Name Full
			(SEQ ID NO. 51)
CH10_MOUSE	52	Hspe1	10 KD heat shock protein, mitochondrial (hsp10) (10K chaperonin) mouse (SEQ ID NO. 52)
CH60_CAEEL	53	hsp60	Chaperonin homolog hsp60 precursor (heat shock protein 60) (hsp-60) (SEQ ID NO. 53)
DEHUE2	54	ALDH2	aldehyde dehydrogenase (NAD+) 2 precursor, mitochondrial (SEQ ID NO. 54)
DEHUE	55	GLUD1	glutamate dehydrogenase (NAD(P)+) precursor (SEQ ID NO. 55)
DEHULP	56	DLD	Dihydrolipoamide dehydrogenase precursor (SEQ ID NO. 56)
DEHUPA	57	PDHA1	pyruvate dehydrogenase (lipoamide) alpha chain precursor (SEQ ID NO. 57)
DEHUPB	58	PDHB	pyruvate dehydrogenase (lipoamide) beta chain precursor (SEQ ID NO. 58)
DEHUPT	59	PDHA2	pyruvate dehydrogenase (lipoamide) alpha chain precursor, testis-specific (E1) (SEQ ID NO. 59)
DEHUXA	60	BCKDH	3-methyl-2-oxobutanoate dehydrogenase (lipoamide) alpha chain precursor (SEQ ID NO. 60)
DEMSMM	61	Mor1	malate dehydrogenase precursor, mitochondrial (SEQ ID NO. 61)
DSHUN	62	SOD2	superoxide dismutase (Mn) precursor (SEQ ID NO. 62)
ECHM_HUMAN	63	ECHS1	enoyl-CoA hydratase, mitochondrial (short chain enoyl-CoA hydratase (SCEH) (SEQ ID NO. 63)
GABT_HUMAN	64	ABAT	4-aminobutyrate aminotransferase, mitochondrial precursor (gamma-amino-N-butyrate-transaminase) (GABA transaminase) (SEQ ID NO. 64)
GCDH_HUMAN	65	GCDH	glutaryl-CoA dehydrogenase precursor (GCD) – human (SEQ ID NO. 65)

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GCDH_MOUSE	66	Gcdh	Glutaryl-CoA dehydrogenase precursor (GCD) – mouse (SEQ ID NO. 66)
HCD1_CAEEL	67	-	Probable 3-hydroxyacyl-CoA dehydrogenase F54C8.1 (SEQ ID NO. 67)
HCD2_CAEEL	68	-	Probable 3-hydroxyacyl-CoA dehydrogenase B0272.3 (SEQ ID NO. 68)
HHMS60	69	Hsp60	heat shock protein 60 precursor (SEQ ID NO. 69)
HMGL_MOUSE	70	Hmgcl	hydroxymethylglutaryl-CoA lyase precursor (HG-CoA lyase) (HL) (3-hydroxy-3-methylglutarate-CoA lyase) (SEQ ID NO. 70)
I48884	71	-	2-oxoglutarate dehydrogenase E1 component (fragment) (SEQ ID NO. 71)
I48966	72	Aldh2	aldehyde dehydrogenase (NAD+) 2 precursor, mitochondrial (SEQ ID NO. 72)
I49605	73	Acads	Acyl-CoA dehydrogenase, short-chain specific precursor (SCAD) (butyryl-CoA dehydrogenase) (SEQ ID NO. 73)
I52240	74	ACAD	acyl-CoA dehydrogenase precursor, medium-chain-specific (SEQ ID NO. 74)
I55465	75	PDK1	pyruvate dehydrogenase kinase isoform 1 – human (SEQ ID NO. 75)
I57023	76	Sod2	superoxide dismutase (Mn) precursor (SEQ ID NO. 76)
I70159	77	PDK2	Pyruvate dehydrogenase kinase isoform 2 – human (SEQ ID NO. 77)
I70160	78	PDK3	pyruvate dehydrogenase kinase isoform 3 – human (SEQ ID NO. 78)
JC2108	79	HADH	long-chain-fatty-acid beta-oxidation multienzyme complex alpha chain precursor, mitochondrial (SEQ ID NO. 79)

<b>MITOP Designation</b>	<b>SEQ. ID. NO.</b>	<b>Gene Name</b>	<b>Gene Name Full</b>
JC2109	80	HADH	long-chain-fatty-acid beta-oxidation multienzyme complex beta chain precursor, mitochondrial (SEQ ID NO. 80)
JC2460	81	PC	pyruvate carboxylase precursor (SEQ ID NO. 81)
JC4879	82	SCHAD	3-hydroxyacyl-CoA dehydrogenase, short chain-specific, precursor (SEQ ID NO. 82)
KIHUA3	83	AK3	nucleoside-triphosphate--adenylate kinase 3 (SEQ ID NO. 83)
M2GD_HUMAN	84	DMGD	Dimethylglycine dehydrogenase, mitochondrial precursor (ME2GLYDH) – human (SEQ ID NO. 84)
MDHM_HUMA	85	MDH2	malate dehydrogenase mitochondrial precursor (fragment) (SEQ ID NO. 85)
O75439	86	PMPC	mitochondrial processing peptidase beta subunit precursor (beta-MPP) (P-52) (SEQ ID NO. 86)
ODO1_MOUSE	87	Ogdh	2-oxoglutarate dehydrogenase E1 component (alpha-ketoglutarate dehydrogenase) (fragment) (SEQ ID NO. 87)
ODPA_CAEEL	88	-	Probable pyruvate dehydrogenase E1 component, alpha subunit precursor (PDHE1-a) (SEQ ID NO. 88)
OWHU	89	OTC	ornithine carbamoyltransferase precursor (SEQ ID NO. 89)
OWMS	90	Otc	ornithine carbamoyltransferase precursor (SEQ ID NO. 90)
P21549	91	AGXT	alanine—glyoxylate aminotransferase (SEQ ID NO. 91)
PUT2_HUMAN	92	ALDH4	Delta-1-pyrroline-5-carboxylate dehydrogenase precursor (P5C dehydrogenase) (SEQ ID NO. 92)
Q0140	93	VAR1	VAR1 – mitochondrial ribosomal protein

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			(SEQ ID NO. 93)
Q10713	94	KIAA0123	mitochondrial processing peptidase alpha subunit precursor (alpha-MPP) (P-55) (HA1523) (SEQ ID NO. 94)
Q16654	95	PDK4	pyruvate dehydrogenase kinase isoform 4 – human (SEQ ID NO. 95)
ROHU	96	TST	thiosulfate sulfurtransferase (SEQ ID NO. 96)
S01174	97	Got2	aspartate transaminase precursor, mitochondrial (SEQ ID NO. 97)
S08680	98	Mut	methylmalonyl-CoA mutase alpha chain precursor (SEQ ID NO. 98)
S13025	99	nuo-40	NADH dehydrogenase (ubiquinone) 40K chain (SEQ ID NO. 99)
S13048	100	cyt	cytochrome c (SEQ ID NO. 100)
S16239	101	Glud	glutamate dehydrogenase (NAD(P)+) precursor (SEQ ID NO. 101)
S23506	102	Pdha1	pyruvate dehydrogenase (lipoamide) (SEQ ID NO. 102)
S25665	103	DLAT_h	dihydrolipoamide S-acetyltransferase heart - human (fragment) (SEQ ID NO. 103)
S26984	104	-	probable DNA-directed RNA polymerase - mitochondrion plasmid maranhar (SGC3) (SEQ ID NO. 104)
S32482	105	ETFB	electron transfer flavoprotein beta chain (SEQ ID NO. 105)
S38770	106	Dci	3,2-trans-enoyl-CoA isomerase, mitochondrial precursor (dodecenoyl-CoA delta-isomerase) (SEQ ID NO. 106)
S39807	107	Bckdhb	3-methyl-2-oxobutanoate dehydrogenase (lipoamide) beta chain (SEQ ID NO. 107)
S40622	108	MUT	methylmalonyl-CoA mutase

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			precursor (MCM) (SEQ ID NO. 108)
S41006	109	-	hypothetical protein t05g5.6 (SEQ ID NO. 109)
S41563	110	cit-1	citrate (si)-synthase, mitochondrial (SEQ ID NO. 110)
S42366	111	PRSS15	Lon proteinase homolog (SEQ ID NO. 111)
S42370	112	-	citrate synthase homolog (SEQ ID NO. 112)
S47532	113	HSPE1	heat shock protein 10 (SEQ ID NO. 113)
S53351	114	ME2.1	malate dehydrogenase (oxaloacetate-decarboxylating) (NADP+) precursor, mitochondrial (SEQ ID NO. 114)
S60028	115	Fdxr	ferredoxin--NADP+ reductase precursor (SEQ ID NO. 115)
S65760	116	Dbt	dihydrolipoamide transacylase precursor (SEQ ID NO. 116)
S71881	117	Bckdha	branched chain alpha-ketoacid dehydrogenase chain E1-alpha precursor (SEQ ID NO. 117)
SCOT_HUMA	118	OXCT	Succinyl-CoA:3-ketoacid-coenzyme A transferase precursor (succinyl CoA:3-oxoacid CoA-transferase) (OXCT) (SEQ ID NO. 118)
SODM_CAEEL	119	sod-2	Superoxide dismutase precursor (Mn) (SEQ ID NO. 119)
SODN_CAEEL	120	sod-3	Superoxide dismutase precursor (Mn) (SEQ ID NO. 120)
SYHUAЕ	121	ALAS2	5-aminolevulinate synthase 2 (SEQ ID NO. 121)
SYHUAL	122	ALAS1	5-aminolevulinate synthase 1 precursor (SEQ ID NO. 122)
SYLM_HUMAN	123	KIAA0028	Probable leucyl-TrNA synthetase, mitochondrial precursor (Leucine--tRNA ligase) (Leurs) (KIAA0028) (SEQ ID NO. 123)



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SYMSAL	124	Alas2	5-aminolevulinate synthase mitochondrial precursor (erythroid- specific) (ALAS-E) (SEQ ID NO. 124)
SYNCLM	125	leu-5	leucine--tRNA ligase precursor, mitochondrial (SEQ ID NO. 125)
SYNCYT	126	cyt-18	tyrosine--tRNA ligase precursor, mitochondrial (SEQ ID NO. 126)
SYWM_CAEEL	127	-	Probable tryptophanyl-tRNA synthetase, mitochondrial (tryptophan--tRNA ligase) (TRPRS) (SEQ ID NO. 127)
THTR_MOUSE	128	Tst	thiosulfate sulfurtransferase (SEQ ID NO. 128)
U80034	129	MIPEP	mitochondrial intermediate peptidase (SEQ ID NO. 129)
U82328	130	PDX1	pyruvate dehydrogenase complex protein X subunit precursor (SEQ ID NO. 130)
XNHU DM	131	GOT2	aspartate transaminase precursor, mitochondrial (SEQ ID NO. 131)
XNHUO	132	OAT	ornithine--oxo-acid transaminase precursor (SEQ ID NO. 132)
XNHUSP	133	AGXT	serine--pyruvate aminotransferase (SPT) (alanine--glyoxylate aminotransferase) (AGT) (SEQ ID NO. 133)
XNMSO	134	Oat	ornithine--oxo-acid transaminase precursor (SEQ ID NO. 134)
XXHU	135	DLAT	dihydrolipoamide S- acetyltransferase precursor (fragment) (SEQ ID NO. 135)
YAL044c	136	GCV3	GCV3 - glycine decarboxylase, subunit H (SEQ ID NO. 136)
YBL022c	137	PIM1	PIM1 - ATP-dependent protease, mitochondrial (SEQ ID NO. 137)
YBL038w	138	MRPL16	MRPL16 -- ribosomal protein of the

MITOP Designation	SEQ. ID. NO.	Gene Name	Gene Name Full
			large subunit, mitochondrial (SEQ ID NO. 138)
YBL080c	139	PET112	PET112 - required to maintain rho+ mitochondrial DNA (SEQ ID NO. 139)
YBL090w	140	MRP21	MRP21 - Mitochondrial ribosomal protein (SEQ ID NO. 140)
YBR120c	141	CBP6	CBP6 - apo-cytochrome B pre-mRNA processing protein (SEQ ID NO. 141)
YBR122c	142	MRPL36	MRPL36 - ribosomal protein YmL36 precursor, mitochondrial (SEQ ID NO. 142)
YBR146w	143	MRPS9	MRPS9 - ribosomal protein S9 precursor, mitochondrial (SEQ ID NO. 143)
YBR221c	144	PDB1	PDB1 - pyruvate dehydrogenase (lipoamide) beta chain precursor
YBR227c	145	MCX1	MCX1 - ClpX homologue in mitochondria
YBR251w	146	MRPS5	MRPS5 - ribosomal protein S5, mitochondrial
YBR268w	147	MRPL37	MRPL37 - ribosomal protein YmL37, mitochondrial
YBR282w	148	MRPL27	MRPL27 - ribosomal protein YmL27 precursor, mitochondrial
YCR003w	149	MRPL32	MRPL32 - ribosomal protein YmL32, mitochondrial
YCR024c	150	-	asn-tRNA synthetase, mitochondrial
YCR028c-a	151	RIM1	RIM1 - ssDNA-binding protein, mitochondrial
YCR046c	152	IMG1	IMG1 - ribosomal protein, mitochondrial
YDL202w	153	MRPL11	MRPL11 - ribosomal protein of the large subunit, mitochondrial
YDR148c	154	KGD2	KGD2 - 2-oxoglutarate dehydrogenase complex E2 component
YDR194c	155	MSS116	MSS116 - RNA helicase of the DEAD box family, mitochondrial
YDR462w	156	MRPL28	MRPL28 - ribosomal protein of the large subunit (YmL28), mitochondrial

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YFL018c	157	LPD1	LPD1 - dihydrolipoamide dehydrogenase precursor
YGR244c	158	LSC2	succinate-CoA ligase beta subunit
YHR008c	159	SOD2	SOD2 - superoxide dismutase (Mn) precursor, mitochondrial
YIL070c	160	MAM33	MAM33 - mitochondrial acidic matrix protein
YJL096w	161	MRPL49	MRPL49 - ribosomal protein YmL49, mitochondrial
YJR113c	162	RSM7	RSM7 - similarity to bacterial, chloroplast and mitochondrial ribosomal protein S7
YKL040c	163	NFU1	NFU1 - iron homeostasis
YLL027w	164	ISA1	ISA1 - mitochondrial protein required for normal iron metabolism
YLR059c	165	REX2	REX2 - putative 3'-5' exonuclease
YML110c	166	COQ5	COQ5 - ubiquinone biosynthesis, methyltransferase
YMR062c	167	ECM40	ECM40 - acetylornithine acetyltransferase
YMR072w	168	ABF2	ABF2 - high mobility group protein
YOL095c	169	HMI1	HMI1 - mitochondrial DNA helicase
YOR040w	170	GLO4	GLO4 - glyoxalase II (hydroxyacylglutathione hydrolase)
YOR142w	171	LSC1	LSC1 - succinate-CoA ligase alpha subunit
YPL118w	172	MRP51	MRP51 - strong similarity to S.kluyveri hypothetical protein
YPL135w	173	ISU1	ISU1 - protein with similarity to iron-sulfur cluster nitrogen fixation proteins
YPL252c	174	YAH1	YAH1 - similarity to adrenodoxin and ferredoxin
YPL262w	175	FUM1	FUM1 - fumarate hydratase
YPR047w	176	MSF1	MSF1 - phenylalanine--tRNA ligase alpha chain, mitochondrial
YPR067w	177	ISA2	ISA2 - mitochondrial protein required for iron metabolism